useful information, or would it be better to arrange for it to be carried out at a specialised centre so as to optimise the diagnostic yield? Each *x*-ray exposure may constitute a very small risk, but many small risks constitute a real danger.

Clinicians are consumers of resources, but frequently they seem to be little aware of the financial consequences of their actions. Even more important, they should be aware of the risks to which they expose their patients in mental and physical terms, in every sense. Patients have a right to be concerned about the risks to which they may be subjected, and doctors have a duty to have sufficient knowledge both to keep those risks to a minimum and to explain them to the patient. In *x*-ray examinations of children the principle of Primum non nocere must be paramount.

[1] SI conversion: 1 rad (old unit) $10^{-2}$ gray (SI unit)
1 rem $10^{-2}$ sievert
1 millirem 10 $\mu$Sv

# Strategic policy monitoring at DHSS

Last year the Commons Select Committee on Social Services turned its attention to the Government's spending plans for the health and personal social services. It produced a report[1] that was sharply critical of the Department of Health and Social Security's ability to formulate and evaluate strategic policies across the range of services for which it has responsibility. In a spirited reply[2] the DHSS pointed out that the committee had failed to ask for information about its machinery for strategic policy making and that had it done so it would have learnt of the existence of several policy-orientated groups and committees within the DHSS and would have been told about the central features of the department's strategic plans.

Now the select committee has returned to the fray, albeit in less strident tones, in a new report[3] on public expenditure in the social services. The committee acknowledged the changes that have been made in the department's planning machinery since its previous report, and it particularly welcomed the setting up of the new Policy Strategy Unit. Even so, the committee argued that any improvements in the machinery for strategic policy making will not necessarily produce more coherent or consistent policies unless those who operate the machinery are more specific about their objectives and more rigorous in monitoring the impact of new policies.

An illustration of the committee's argument is found in its discussion of the effects of changes in the level of spending on particular services, programmes, and client groups. An increase in expenditure might lead to an improvement in the quality of a service, but it might equally result in an expansion in the volume of the service, an extension in its coverage, or an increase in its cost. The committee argued that these effects were very different in their social impact and that policy makers should therefore be clear about their aims when deciding to increase expenditure and should subsequently be able to assess the extent to which those aims had been fulfilled. Accordingly, the committee recommended that every effort should be made to find new ways of measuring the quality of care, the savings arising from improvements in efficiency, and the effectiveness of programmes in meeting established policy goals.

Understandably, however, the select committee's enthusiasm for what it called "strategic consistency and coherence" is not shared unreservedly by the DHSS. In its earlier rejoinder to the committee,[3] the department pointed out that it is often unrealistic and impracticable to specify policy objectives or to measure their impact with the precision demanded by the committee. Decisions which have to be taken on the basis of less-than-perfect information may easily be criticised later with the benefit of hindsight. Yet the department also seems deliberately to be taking steps that will impair its ability to behave in ways expected by the committee. The DHSS's failure to specify priorities in resource terms in its recent handbook on policies,[4] its determination to shift some of the emphasis in making policy away from the centre towards the regions and districts, and its proposals[5] to curtail the available statistical information about health and health services will make it more difficult for the department to be accountable for its policies in the way that the committee would wish.

In this sense, the continuing dialogue between the Select Committee on Social Services and the DHSS provides a fascinating illustration of the tension between those who favour a comprehensive rational approach to making policy and those who favour a judicious blend of rationality and pragmatism. The new report from the committee resolves nothing but it plays a sharp volley back to the Secretary of State's court by asking him to identify precisely the guidelines by which the committee and others can monitor the progress being made towards the Government's policy objectives. Doctors will await with interest his return stroke.


[1] Social Services Committee. *Third report from the Social Services Committee 1979-80: the Government's White Papers on public expenditure: the social services.* London: HMSO, 1980. (HC 702.)

[2] Department of Health and Social Security. *Reply by the Government to the third report from the Social Services Committee, session 1979-80.* London: HMSO. (Cmnd 8086.)

[3] Social Services Committee. *Third report from the Social Services Committee 1980-81: public expenditure on the social services.* London: HMSO, 1981. (HC 324.)

[4] Department of Health and Social Security. *Care in action. A handbook of policies and priorities for the health and personal social services in England.* London: HMSO, 1981.

[5] Lord President of the Council. Privy Council Office. *Government statistical services.* London: HMSO, 1981. (Cmnd 8236.)


# Alpha$_1$-antitrypsin deficiency and liver disease

Alpha$_1$-antitrypsin, a glycoprotein with a molecular weight of 54 000 which is synthesised and catabolised in the liver, is a major serum protease inhibitor but its physiological function is still unknown.

The genetics of alpha$_1$-antitrypsin deficiency are well worked out, with at least 26 alleles identifiable by starch-gel electrophoresis or by isoelectric focusing. The protease inhibitor (Pi) alleles are labelled alphabetically, with the electrophoretically slowest being designated Z, the medium M, and the fastest F. The most common allele is PiM, its frequency being at least 0·87.[1] Deficiency of alpha$_1$-antitrypsin is usually associated with the phenotype PiZZ and sometimes PiSZ. Patients have been described in whom alpha$_1$-antitrypsin is virtually undetectable and no electrophoretic pattern is available; the allele in this case is called $Pi^{null}$.[2]

The inheritance of the alleles follows an autosomal codominant pattern. Histological examination of the liver of individuals who are homozygous or heterozygous for the phenotype PiZ shows that the hepatocytes contain character-

istic globules of an amorphous material which are resistant to diastase and positive for periodic-acid-Schiff. Immunofluorescence studies have shown that these globules are immunologically similar to alpha$_1$-antitrypsin; they may be the result of accumulation of a precursor of alpha$_1$-antitrypsin which cannot be released from the hepatocytes possibly because of a modification in its structure. The material extracted from the globules contains no sialic acid—part of the circulating alpha$_1$-antitrypsin molecule.[3]

The association between deficiency of alpha$_1$-antitrypsin and childhood cirrhosis was first described in 1976[4] and since confirmed in numerous studies in both children and adults. In childhood, liver disease associated with alpha$_1$-antitrypsin deficiency usually presents in the first four months of life as an acute hepatitis with conjugated hyperbilirubinaemia and often follows directly neonatal physiological jaundice. The characteristic periodic-acid-Schiff-positive globules within the hepatocytes are rarely seen before 12 weeks of age, despite florid liver damage.[5] The clinical severity of the hepatitis is variable. About one-quarter of the children presenting with neonatal hepatitis die from cirrhosis by the second decade of life, one-quarter have cirrhosis, one-quarter have persistently abnormal liver function values, and one-quarter seem to recover completely.[6] Not all PiZ infants develop clinical features of neonatal hepatitis. A comprehensive epidemiological study in Sweden found that 11% of infants with alpha$_1$-antitrypsin deficiency developed prolonged cholestatic jaundice; 6% had subclinical hepatitis in infancy without jaundice; and 35% had minor abnormalities of liver function.[7] Why only some PiZ infants develop liver disease is not known. Possibly the liver is unable to control a damaging process caused by environmental or associated genetic factors, which would have been adequately controlled had normal inhibitors of bacterial, viral, or inflammatory cell proteases been present.

In adults the association between the homozygous ZZ phenotype and liver disease is even less clear, and the incidence of such an association varies considerably.[8 9] Furthermore, there are appreciable differences in the prevalence of hepatic fibrosis, cirrhosis, or hepatocellular carcinoma in different geographical areas.[10–13]

Whether an association exists between liver disease and the heterozygous PiZ state remains uncertain. No association was found when patients were screened by measuring serum levels of alpha$_1$-antitrypsin[13] or by phenotyping the patients.[14 15] In contrast, an association was seen when only patients with the characteristic periodic-acid-Schiff-positive inclusions in the hepatocytes were studied.[16] Hodges *et al*[17] have recently reported the results of a five-year prospective study of liver biopsy specimens from 1055 adults with liver disease. Phenotyping of the 34 patients whose specimens contained characteristic hepatocyte inclusions showed phenotype MZ in 25 of them. The other phenotypes found were ZZ, SZ, MS, and MM. Twenty-one per cent of patients with cryptogenic cirrhosis and 20·5% of those with chronic active hepatitis negative for hepatitis B surface antigen had phenotype MZ, whereas this was found in only 3·5% of patients with alcoholic cirrhosis and 2·6% of those with other types of cirrhosis. Hodges *et al*[17] also suggested that the patients with chronic active hepatitis and phenotype MZ might have distinctive clinical features.

The mechanism of liver damage in such patients remains to be explained. It does not seem to be correlated with low serum activities of alpha$_1$-antitrypsin; 42% of the heterozygous patients with chronic active hepatitis or cryptogenic cirrhosis reported by Hodges *et al* had values within the normal range at presentation. Nor is the damage likely to result from accumulation of alpha$_1$-antitrypsin in the liver, since patients without liver damage have periodic-acid-Schiff-positive globules. Further studies from different geographical areas are needed to find out whether there is a true association between the PiMZ phenotype and liver disease or whether liver damage is secondary to unknown associated environmental or genetic factors.


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# Treatment of seasonal and perennial rhinitis

Hyperreactivity of the nasal mucosa causes a range of disorders whose main symptoms are sneezing, itching, rhinorrhoea, nasal congestion, and blockage. These symptoms are usually labelled as seasonal or perennial allergic rhinitis when there is a recognised provoking antigen and as vasomotor (or non-allergic) rhinitis when there is not. Since some patients with seasonal allergic rhinitis (hay fever) may have nasal symptoms all the year these disorders may prove to be a continuum rather than separate diseases.[1]

Allergic rhinitis, whether seasonal or perennial, is mainly due to a type 1 allergic reaction. Specific IgE immunoglobulins become attached to the surface of the mast cells, and when the patient is re-exposed to the antigen these cells release histamine and other chemical mediators, causing sneezing, nasal itching, rhinorrhoea, and nasal congestion. Effective treatment depends on either preventing the release of mediators or blocking their pharmacological effects.

In contrast, the mechanisms underlying vasomotor and